Humphry

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THE ANATOMY OF SPINA BIFIDA. By Professor Humphry.

(*Delivered as a Museum-Lecture in the University of Cambridge, and taken, with a few Alterations and Additions, from the "Lancet," March* 25, 1885.)

Gentlemen,—Finding considerable difference of opinion among surgeons respecting the anatomy of spina bifida, and especially, at which I was surprised, respecting the usual relations of the spinal cord and the spinal nerves to the sac, I have recently examined the specimens of this affection in most of the pathological collections of London, as well as some recent specimens which have been kindly submitted to me by Dr Goodhart and Mr Stewart. The chief points are, however, sufficiently demonstrated in the specimens before you, most of which were dissected and placed in the museum of the University by myself a good many years ago.

The name "spina bifida" is derived from one, and that not the most important, feature of the affection—viz., the separation or divergence, and in some instances the dwarfing, of the vertebral arches. This is associated with, and probably caused by, the protrusion from the vertebral canal of a sac containing a thin colourless fluid situated in the middle line, and of variable size. The affection is most common in the sacral, or the lumbo-sacral, region. The sac is more or less covered by skin, rarely altogether, but commonly to some extent of its circumference; the middle part being usually covered, rather formed, by a thin, translucent, bluish-red, more or less vascular, membrane; though in some instances this middle part has the appearance of a cicatrix. The thin, translucent, or cicatricial middle part, sometimes called the "Umbilicus," is continuous with the surrounding skin, the line of demarcation between the two being pretty clearly defined. The sac is commonly circular in outline, and its upper and lower borders afford some criterion of the extent in the spinal column to which the vertebral arches are deflected and the vertebral canal is opened. Still, the sac not unfrequently overlaps the unaffected arches above and below, and it usually overlies and extends beyond the everted arches at the sides. Dissection in most instances shows the spinal cord turning beneath the lowest entire vertebral arch—that is, the arch immediately above the spina bifida—and passing into some part of the wall of the sac. In some cases the cord runs, free, through the upper part of the sac to its middle, where the wall is composed of the thin membrane just mentioned. In other instances, immediately upon reaching the sac, the cord is blended with the wall of the sac, and requires a little dissection for its demonstration. The nerves, traced from the sacral, or the lumbar, intervertebral foramina, do not form a cauda equina in the usual manner, but are directed transversely, or nearly so, towards the same membranous part of the sac as the spinal cord; and, like the spinal cord, they are, in some instances, seen, in their course to this

part, lying free in the sac, whereas in others they lie, in their whole length, in the wall of the sac. In a third set of cases they present intermediate conditions, being partially free in the sac and partially embedded in its wall; or they are more or less united together by membranous bands which may be regarded as derivatives from the inner layer of the wall of the sac; and it is probable that the compartments, and even the cysts, occasionally found in connection with spina bifida sacs, may be thus produced. Thus, both cord and nerves pass to the sac, being stretched or lengthened in proportion to the size of the sac; and they are more or less blended with the thin membranous hinder part of it. In this specimen—at the lumbar and upper sacral region—the cord is seen entering the sac, traversing the upper part of its wall, blended with the hinder thin part, and then reappearing at the lower part, where something of a cauda equina is formed by the junction of the nerves of the two sides with the cord, and the median filament extends onwards towards the coccyx. The cord therefore in this case may be traced, and the anterior spinal artery is visible upon its surface, along the whole of the interior of the back of the sac, and from the sac into the sacrum; and the several lumbar nerves, from the two sides, pass on the wall of the sac to the cord. There was hydrocephalus in this case; and the central spinal canal, which communicated above with the distended cerebral ventricles, is dilated down to the spina bifida.

The nerve-roots and their ganglia in cases of spina bifida are not infrequently situated within the vertebral canal; and in some of the specimens before you, two or three adjacent ganglia are seen to be approximated and even joined together, and contained in one sheath of dura mater. This approximation and fusion are probably a result of the traction exerted upon the nerves in their passage upon the wall of the sac.[1]

The cavity of the spina bifida, which, as we have seen, is, in some cases, traversed by nerves and partitioned by membranous septa, looks, in many specimens, as if it were continuous with the arachnoid sac, and as if it were lined by the arachnoid membrane. In some this is actually the case. It is so in the dorsi-lumbar specimen I have just shown you. The sac of the spina bifida is here formed by a dilatation of the anterior part of the spinal arachnoid cavity, the posterior part of that cavity, the part behind the nerve roots, being obliterated, or not having been formed. But a careful and close examination shows, in some at least, perhaps in most of those in the sacral or sacro-lumbar region, that this is not the case. The parietal and visceral surfaces of the arachnoid in the spinal canal, near to the sac or at its orifice, may be seen to be adherent; or the epithelial lining is reflected from the one to the other, so that the arachnoid membrane is not traceable into the spina bifida, and the spinal arachnoid cavity is shut off from the cavity of the spina bifida. The sac of the spina bifida accordingly appears to be formed in the subarachnoid space, and the lining of the

[1] This fusion of the ganglia was pointed out by Dr Fisher Downing, Professor of Medicine in this University (*London and Ed. Phil. Mag.*, vol. x. 516, 1837).

sac, as well as the coverings of the cord and of the nerves, and the more or less complete membranous septa, where these exist, must in that case be formed by the subarachnoid, or pia mater, tissue. It is not always easy to determine, even on dissection, whether the sac of the spina bifida is thus separate from the arachnoid cavity; but where it is so the fluid in the sac, and, in like manner, any fluid injected into the sac will not readily pass into and along the arachnoid cavity of the spinal canal and the skull, though it may permeate the thin membrane which forms the wall of the sac, and so reach the arachnoid cavity or the loose subarachnoid tissue of the cord, and thus extend along the cord to the brain.

The specimen of lumbo-sacral spina bifida which I have here, entrusted to me by Dr Goodhart, and which has been recently dissected for me by Dr Anningson, shows well many points of the anatomy of this malformation. The sac is of about the size of an orange, with the usual cicatricial tissue at the middle of its wall and with skin over the circumference. The skin and fat were easily removed near the spine, but are adherent to the subjacent layers of the sac near the middle, and ultimately blend with them in the cicatricial tissue. A tough structure, apparently the lateral halves of the interspinous ligament cleft like the spinous processes, was found, beneath the skin and fat, on the sides of the sac. It was connected laterally with the recurved and shortened vertebral arches and spinous processes. The dura mater of the cord expands over the sac, forming a thick covering pierced by the spinal nerves at the sides and blended with the other structures at the border of the cicatricial tissue. Beneath the dura mater is a double layer—visceral or spinal, and peripheral or subdural—of the posterior arachnoid (the arachnoid, that is, behind the nerve roots and the ligamentum denticulatum). These layers, though in close contact, were easily separated, the posterior arachnoid cavity being thus displayed. Like the other components of the sac, they are adherent and blended in the cicatricial tissue, and they are also closely adherent at the junction of the spina bifida with the spinal canal—at the level, that is, of the lowest complete vertebral arch; so that the posterior arachnoid cavity of the sac (in the wall of the sac) is quite shut off from the posterior arachnoid cavity of the rest of the spine. The visceral and peripheral surfaces of the anterior arachnoid (the arachnoid that is in front of the spinal nerves and the ligamentum denticulatum) were non-adherent; and the anterior arachnoid cavity was quite free along the whole length of the vertebral canal, including the spina bifida. The cavity of the sac is formed altogether in the anterior subarachnoid space, is entirely lined by subarachnoid or pia mater tissue, and is partially divided by imperfect septa of that tissue, projected into it by the spinal cord and nerves. A thick nerve-bundle, consisting of the spinal cord, with surrounding lumbar and upper sacral nerves, and forming, as it were, a retroverted cauda equina, turns beneath the last vertebral arch, leaving the bodies of the vertebræ, the anterior dura mater, and the anterior arachnoid. Thus, invested with pia mater or subarachnoid tissue only, it runs for a short space free in

the sac to the cicatricial tissue of the hinder wall of the sac; here the component cord and nerves, with their investing pia mater, are lost in the cicatricial tissue. The spinal nerves pass from the intervertebral foramina in regular order and pierce the dura mater separately all the way down from the head to the coccyx. The lower lumbar nerves having entered the dura mater, are, as we have seen, directed downwards, accompanying the cord to the back of the sac. The upper two sacral nerves, piercing the wall of the sac, take a more transverse course to the same point; and their ganglia, like those of the lumbar nerves, are outside the cavity of the arachnoid, and invested with sheaths of the dura mater. The nerve roots of the lower sacral nerves also pierce the dura mater, then run between the arachnoid layers in the wall of the sac—that is, in the posterior arachnoid cavity of the wall of the sac,—and so reach the cicatricial tissue; and the ganglia of their posterior roots lie in this posterior arachnoid cavity. Thus the wall of the sac is formed by the dura mater, the arachnoid, and the outer layers of the pia mater, together with the spinal cord and nerves; and the cavity of the sac is formed in the anterior subarachnoid tissue, and is lined, probably, by the endothelium of that tissue. Along the fore part of the vertebral canal the three coats, with the anterior arachnoid cavity, are undisturbed, and retain their normal relations with the bodies of the vertebræ, except that, as just said, the cavity is formed in the anterior subarachnoid, or pia mater, tissue, and a layer of that tissue is therefore expanded over the cavity; whereas at the hinder part of the vertebral canal, the three coats with the posterior arachnoid cavity, together with the nerves and the spinal cord, are stretched out to form the bulging wall of the sac, are more or less adherent to one another, and are all, as well as the cutaneous coverings, lost in the cicatricial tissue of the hindmost part of the sac. This specimen may be taken as illustrating the usual anatomy of a spina bifida, an accumulation of fluid in a cavity formed in the anterior subarachnoid or pia mater, with all the structures posterior to it—namely, subarachnoid tissue, spinal cord and nerves, posterior arachnoid membrane and cavity, dura mater, vertebral arches, subcutaneous and cutaneous tissues—stretched over it, and more or less imperfectly formed.

It is evident from the above anatomical considerations that the failure of development upon which spina bifida depends must occur at an early period of fœtal life, in most instances before the spinal cord has been segmented from the epiblastic, or epithelial, layer of the embryo from which it is developed. It accordingly remains adherent to that external epiblastic covering; the imperfectly developed tissues of the cord and of the nerves passing to it, together with this covering, form the membranous wall of the sac; and the various structures which should be produced between the cord and the epiblast are to a greater or less extent abortive. The failure may even take place, it is believed by some usually to do so, before the central spinal canal has been closed; or the recently closed canal may give way and be reopened by fluid accumulating in its interior; and in such case the

median posterior exposed surface of the sac must correspond with the epiblastic tissue which should have formed the epithelial lining of the central canal.[1] This often cannot easily be determined, for the central canal becomes occluded where the cord joins the membranous wall of the sac, in the lower regions of the vertebral column at any rate; and its precise relations to the sac cannot therefore be made out. It was so in the specimen we have just seen, when there were hydrocephalus and dilatation of the central canal down to the spina bifida. The canal in this case could easily be traced down to the spina bifida, but no further; and it was impossible to say positively whether the structure which represented it was continued into the cord lining the sac of the spina bifida, or whether it was continued upon the external surface of the sac, though the appearances are in favour of the former supposition.

Associated with the failure of segmentation and development of the several structures between the cord and the epiblast is the accumulation of fluid, or dropsy, by the pressure of which the sac is formed, the vertebral arches are recurved or stunted, the nerves and the spinal cord are elongated, and the tumour is produced. It is possible that the dropsical condition, due perhaps to something of an inflammatory process, may be the prime agent, and that by its pressure it has prevented the segmentation of the tissues from the epiblast, as well as induced the other changes mentioned; or the dropsy may be due to a want of resistance consequent on the non-segmentation and arrested development of those tissues—that is to say, it may be cause or effect. The balance of opinion and of probability is perhaps in favour of the former view. Further, when several vertebræ above the sacrum are affected, the fore part of the spine, composed of the bodies of the vertebræ and the intervertebral substances, being unsupported by the vertebral arches and articulating processes, which are abortive or deflected, may bend backwards or forwards. In this specimen, in the loins, it is bent backwards; in this specimen, in the neck, it is bent forwards, so that the chin rests on the sternum; and in this third example, also in the neck, it is bent backwards, so that the occiput lies on the back. These several defects in the vertebral column, including the open state of the vertebral canal with the separation, eversion, or stunting of the vertebral arches, are all consequences of the arrested development and the imperfect segmentation and imperfect formation of the various structures in the median line, and of the dropsical accumulation associated with them. The greater frequency of this failure and of the presence of spina bifida in the lower —the sacral and lumbo-sacral—parts of the spine, is probably owing to the fact that the closing of the primitive furrow, which is a little dilated at this part, and other developmental phenomena take place here rather later than in the upper regions; and they are therefore more likely to be interfered with or to be imperfect in this situation. A trace of this imperfection in the otherwise well-formed body is often

[1] See Contribution to the Study of Spina Bifida by Professor Cleland, *Journal of Anatomy and Physiology*, vol. xvii. p. 257.

to be seen in the smallness or incompleteness of the arch of the upper sacral vertebra, and also in the dimple in the skin over the coccyx.

I have thus described to you the anatomy of spina bifida, as it usually occurs in the sacral or the lumbo-sacral region; and you will have noted that the accumulation of fluid is usually in the subarachnoid space in front of the spinal cord, that the cord and the nerves are stretched backwards and outwards upon the sac, and are there confluent, together with the arachnoid, pia mater, and dura mater, or their representatives, in the thin membrane which forms the hindmost part of the wall of the sac. Moreover, it is to be remarked that the cord, being unsegmented from this thin membrane, and therefore fixed at this spot, and so fixed from a very early period of fœtal life, does not undergo that shifting in relation to the vertebral canal which ordinarily takes place during the later periods of fœtal life, and which is due to a greater proportionate growth of the vertebræ as compared with that of the cord. Hence the spinal cord, instead of terminating, as it does in the normal state, in the upper lumbar region, extends through the lumbar region into the wall of the sac, and may, in some instances, be traced beyond it to the lower part of the sacrum; and, as a sequence, the sacral and lumbar nerves pass transversely towards the cord in the sac, instead of ascending obliquely and forming a cauda equina; indeed, not unfrequently, as in a specimen I have shown you, the nerves traversing the spinal foramina just above the spina bifida take the reverse of their usual course and pass downwards, rather are dragged downwards, to the sac to join the cord. This, I say, is the usual disposition as demonstrated by much the larger number of specimens. In some cases, however, when the failure has occurred at a later period of fœtal life, the segmentation and the formative development of parts have taken place to a greater extent; and the skin, with its epithelium and the subcutaneous tissue, may have been formed in a natural manner all over the sac. This is the most frequent in the neck. In some, as in this lumbo-sacral specimen before shown you, the neural axis is more separated than in the other cases—that is to say, it is less completely fused with the wall of the sac, and forms a distinct cord or band attached along the whole of the inner surface of the hinder wall of the sac. It may be, too, that in a few cases the segmentation and formation of the cord have been still more complete, so that it has become fully separated from the cutaneous and other tissues, and retains its normal place along the bodies of the vertebræ, and the dropsy, with the sac formed by it, is upon the hinder aspect of the cord. The cord would then not pass into the sac, at least would not pass along, and would not be attached to, the hinder bulging wall of the sac. This would constitute a "hydro-meningocele" or "hydrorachis externa" *posterior*, in contradistinction to a "hydrorachis externa" *anterior* in which the cord and nerves enter the sac, and blend with its wall; and both these are to be distinguished from a "hydro-myclocele" or "hydrorachis interna," in which the fluid accumulates in the central spinal canal. Possibly this is so—that the fluid is behind the cord—in a specimen in the museum of St Bartholomew's Hospital. The

specimen is a small spina bifida at the lower part of the sacrum, and the median filament of the cord and the nerves seem to be at the fore part of the sac; but further dissection is necessary to make the anatomy clear. In the College of Surgeons' museum is a specimen from an infant who died three days after the application of a ligature to a cervical spina bifida. The narrow peduncle of the sac passes through between the sixth and seventh cervical spinous processes, which are almost fully developed, and nearly meet in the middle line. The cavity of the sac appears, from the direction of a bristle placed in it, to communicate with the hinder part of the arachnoid cavity, and the cord possibly is not in the least degree implicated, and passes free along the hinder surface of the bodies of the vertebræ. But further dissection is required to make sure of this. It may be, as in an apparently similar case represented in Förster's *Missbildungen*, that the affection consists in a dilatation of the central spinal canal, and that a thin dilated sheet of the hinder part of the cord is prolonged into the wall of the sac—that is to say, it may be an example of "hydrorachis interna." [1] This can only be determined, if in such a specimen it can be determined at all, by removing the dura mater, and making a careful examination of the parts. Whether the sac in this case was covered by skin or not cannot be seen. In cases of cervical spina bifida, occasionally met with, in which the sac has a narrow neck, and is covered by skin, the disposition is probably similar to that in the specimen just referred to. Like it, such cases furnish examples of the slightest degree of the malformation; and the examination of a larger number of specimens of the kind may enable us to judge what inference, as to the disposition of the cord and its relations to the sac, may be drawn from the presence of a covering of skin over the sac—whether, that is to say, the segmentation and development which have produced skin over the surface are associated with a corresponding segmentation and development of the cord sufficient to set it, with its pia mater and arachnoid, free from the sac, or so far free as to allow it to maintain its proper place in contact with the bodies of the vertebræ.

The fluid, as I have already said, appears commonly to be in the subarachnoid space in front of the cord; and the sac must in such cases, I conclude, be lined by an endothelium developed from that of the subarachnoid lymph spaces. In some instances the fluid is in the arachnoid cavity in front of the cord, and then the sac is lined by the endothelium of the arachnoid. In hydrorachis interna, where the sac is formed by a dilatation of the central spinal canal, the epithelial lining of the sac will be continuous with that of the central canal. The fluid may be, though this rarely is so, in the subarachnoid

[1] That is the case in a well-preserved and well-dissected specimen in the Musée Dupuytren in Paris. There is hydrocephalus and a spina bifida sac at the seventh dorsal vertebra. The spinal cord is normal above and below this point. At this point it is bent a little backwards towards the sac, and the hinder part of it is connected with, apparently expanded over, the sac. Some of the nerve fibres, those apparently from the posterior roots, have been exposed by dissection in the wall of the sac; and it is stated that the cavity of the sac is continuous with the canal of the cord.

space, or in the arachnoid cavity *behind* the cord. In these last cases the cord would occupy its normal position upon the bodies of the vertebræ, and the nerves would also have their normal position; whereas in the cases in which the fluid, whether arachnoidal or subarachnoidal, is in *front*, the cord and nerves are separated from the bodies of the vertebræ, and are applied upon and coherent to, or blended with, the wall of the sac. The dura mater is in all instances expanded upon the sac, and traceable to the same extent as the skin, becoming like it, and at about the same place, lost in the homogeneous membrane of the hinder part of the sac. Though I admit that the fluid may be behind the cord, the cord retaining its normal position, I must say that I have not been able to satisfy myself that this was the condition in any one of the many specimens I have examined. In every one, unless the one presently to be mentioned be an exception, in which the anatomy was clear, the cord, or some part of it, and usually the nerves, passed into the sac, and was united with the wall of the sac.

In those cases where the vertebral arches are more extensively deficient, and which are often associated with deficiency of the vault of the skull and with more or less deficiency of the brain, the vertebral canal is usually covered only by a thin membrane, and the spinal cord, with the several superadjacent tissues, is absent or represented by that membrane into which the spinal nerves pass and are lost. In the specimen, however, which I now show you this is not the case. The fœtus is acranial and with spina bifida along the whole spine; the neck is bent sharply forward, so that the back of the head is in contact with the dorsal part of the spine. The cerebral mass is quite rudimentary; and the axial cord, of considerable thickness, passes directly backwards from the floor of the skull, bridging over the concavity of the neck, upon the dorsal vertebræ, and is continued upon the bodies of the vertebræ along the whole length of the lower dorsal, the lumbar, and the sacral portions of the spine. The nerves join it in regular sequence; but in the neck they are crowded together, and the lower cervical ganglia on each side are blended into one mass invested by a common capsule of dura mater. In the upper part of the cord the central spinal canal is wide and open, forming a continuation of the fourth ventricle. It gradually diminishes to the lower dorsal region, where it is closed in as usual; and below this the cord tapers into the median filament, which is continued to the coccyx. The specimen when it came into my hands, many years ago, was in its present condition; and to what extent the spinal central canal was dilated in the upper part, how it was covered in, what kind of covering existed over the spina bifida, and what was the cause of the eversion of the vertebral arches could not be ascertained. All that we can see is that the vertebral arches, without evident diminution of size, are thrown outwards and forwards, the vertebral canal is wide open in its whole length, and the spinal column is bent sharply forwards in the cervical region, and backwards in the dorsal region. The cord and the nerves lie on the bodies of the vertebræ in the sacral, lumbar, and

dorsal regions, and pass, straight over the concavity of the cervical curve, from the dorsal region to the skull; and in this part where it bridges over the curve the central spinal canal is dilated. It is a remarkable case, and it appears to constitute an exception to the rule of the cord being closely connected with the hinder wall of a spina bifida, and to the ordinary manner of formation of spina bifida.

I say nothing about other exceptional varieties of spina bifida, such, for instance, as where there is cleavage of the bodies of the vertebræ, or bony or cartilaginous growths from them into the vertebral canal, and other malformations. I have directed your attention chiefly to the more ordinary forms of spina bifida, the anatomy of which has acquired increased importance from the reintroduction of the treatment of the abnormality by injection. With reference to this treatment, you will understand that it is better to introduce the needle, not in the mesial line, especially in the upper part, where the cord is almost certain to be connected with the wall of the sac, but on one side. Further, you will perceive that the separateness of the cavity of the sac from the cavity of the spinal arachnoid, in most instances, is some security against the direct passage of the injected fluid into the latter cavity, and that the advantage of Morton's fluid, as an injection, probably depends upon the addition of glycerine to the iodine and iodide of potassium lessening the liability to the diffusion of those irritating substances through the delicate lining of the sac into the surrounding subarachnoid tissue.